

**In the Specification:**

Please amend the specification as shown:

Please delete the paragraphs on page 9, line 24 to page 10, line 25 and replace them with the following paragraphs:

**Figure 5.** RNAi-mediated suppression of expanded CAG repeat containing genes.

Expanded CAG repeats are not direct targets for preferential inactivation (A), but a linked SNP can be exploited to generate siRNA that selectively silences mutant ataxin-3 expression (B-F). (A) Schematic of cDNA encoding generalized polyQ-fluorescent protein fusions. Bars indicate regions targeted by siRNAs. HeLa cells co-transfected with Q80-GFP, Q19-RFP and the indicated siRNA. Nuclei are visualized by DAPI staining (blue) in merged images.

(B) Schematic of human *ataxin-3* cDNA with bars indicating regions targeted by siRNAs. The targeted SNP (G987C) is shown in color. In the displayed siRNAs, red or blue bars denote C or G respectively. In this Figure, CAGCAGCAGCAGGGGACCTATCAGGAC is SEQ ID NO:7, and

CAGCAGCAGCAGCGGGACCTATCAGGAC is SEQ ID NO:8. (C) Quantitation of fluorescence in Cos-7 cells transfected with wild type or mutant ataxin-3-GFP expression plasmids and the indicated siRNA. Fluorescence from cells co-transfected with siMiss was set at one. Bars depict mean total fluorescence from three independent experiments +/- standard error of the mean (SEM). (D) Western blot analysis of cells co-transfected with the indicated ataxin-3 expression plasmids (top) and siRNAs (bottom). Appearance of aggregated, mutant ataxin-3 in the stacking gel (seen with siMiss and siG10) is prevented by siRNA inhibition of the mutant allele. (E) Allele specificity is retained in the simulated heterozygous state. Western blot analysis of Cos-7 cells cotransfected with wild-type (atx-3-Q28-GFP) and mutant (atx-Q166) expression plasmids along with the indicated siRNAs. (Mutant ataxin-3 detected with 1C2, an antibody specific for expanded polyQ, and wild-type ataxin-3 detected with anti-ataxin-3 antibody.) (F) Western blot of Cos-7 cells transfected with Atx-3-GFP expression plasmids and plasmids encoding the indicated shRNA. The negative control plasmid, phU6-LacZi, encodes siRNA specific for LacZ. Both normal and

mutant protein were detected with anti-ataxin-3 antibody. Tubulin immunostaining shown as a loading control in panels (D)-(F).

**Figure 6.** Primer sequences (SEQ ID NOS: 11-40, respectively, in order of appearance) for *in vitro* synthesis of siRNAs using T7 polymerase. All primers contain the following T7 promoter sequence at their 3' ends: 5'-TATAGTGAGTCGTATTA-3' (SEQ ID NO:9). The following primer was annealed to all oligos to synthesize siRNAs: 5'-TAATACGACTCACTATAG-3' (SEQ ID NO:10).

Please delete the paragraph on page 12, lines 24-30 and replace it with the following paragraph:

**Figure 12.** Design and targeted sequences of siRNAs (SEQ ID NOS: 42-54, respectively, in order of appearance). Shown are the relative positions and targeted mRNA sequences for each primer used in this study. Mis-siRNA (negative control) does not target TA; com-siRNA targets a sequence present in wild type and mutant TA; wt-siRNA targets only wild type TA; and three mutant-specific siRNAs (Mut A, B, C) preferentially target mutant TA. The pair of GAG codons near the c-terminus of wild type mRNA are shown in underlined gray and black, with one codon deleted in mutant mRNA.